Atrial Septal Defect of the Persistent Ostium Primum Type with Hypoplastic Right Ventricle in a Welsh Pony Foal

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ABSTRACT

Valvular competency of the foramen ovale (patent foramen ovale) is regarded as a common finding in the neonatal foal and usually occurs in isolation. True atrial septal defects appear to be uncommon and are usually associated with other congenital cardiac lesions. The present report describes a case of atrial septal defect type 1 (persistent ostium primum) complicated by hypoplastic right ventricle, and tricuspid dysplasia, in a Welsh Mountain pony foal, and discusses the embryogenesis of the abnormality. A critical review of the literature suggests that atrial septal defects may occur more frequently than they are reported, and that on occasion they may be described erroneously as patent foramen ovale. The clinical significance of uncomplicated discontinuity of the atrial septum is slight, depending upon the size and location of the defect. Complicated atrial septal defects vary in clinical significance according to the nature of the associated defects.

Key words: Equine, cardiac defect, patent foramen ovale, atrial septal defect.

RÉSUMÉ

La présence d'un *foramen ovale* perméable, capable de fonctionner comme une valvule, représente une observation fréquente, chez le poulain nouveau-né; cette anomalie se rencontre ordinairement comme une communication interauriculaire isolée. Les véritables anomalies de la cloison interauriculaire semblent plutôt rares et s'accompagnent ordinairement d'autres lésions cardiaques congénitales. Les auteurs décrivent une anomalie de la cloison interauriculaire du type #1, i.e. l'*ostium primum* persistant, compliquée d'une hypoplasie du ventricule droit et d'une dysplasie de la valvule tricuspide, chez un poulain poney "Welsh Mountain"; ils commentent aussi l'embryogénèse de cette anomalie. Une revue critique de la littérature permet de penser que les véritables anomalies de la cloison interauriculaire se produiraient plus souvent qu'on les rapporte et qu'on les décrirait occasionnellement, erronément, comme un *foramen ovale* perméable. La signification clinique d'une discontinuité non compliquée de la paroi interauriculaire est proportionnelle à ses dimensions et à sa localisation, tandis que celle des véritables anomalies de la cloison interauriculaire varie selon la nature des autres anomalies qui les accompagnent.

Mots clés: équins, anomalie cardiaque, *foramen ovale* perméable, anomalie de la paroi interauriculaire.

INTRODUCTION

Congenital discontinuity of the atrial septum in the form of valvular competency (1) of a potential opening, the foramen ovale (a condition commonly referred to as patent foramen ovale) is generally regarded as a common finding in the neonatal foal (2,3,4). Patency may vary from minute fenestrations of the sleeve of tissue which guards the opening of the foramen in the left atrium, to openings a centimetre or more in diameter. However, the literature contains only four specific references to cases of patent foramen ovale in foals (5,6,7,8). Relatively few reports of true atrial septal defect (ASD) have been found; atrial septal abnormalities other than patent foramen ovale are regarded as rare (3).

Four cases of ASD associated with tricuspid atresia have been reported (6,10,11). Tricuspid atresia has also been associated with patent foramen ovale in the horse (5). Atrial septal defect has been associated with pulmonic stenosis or pulmonary atresia in four cases (7,10,12,13), while in an additional case the defect occurred as a component of a common atrioventricular canal (14). An atrial septal defect, probably of the inferior caval type, has been reported in a foal in association with hypoplastic left heart syndrome (15), and one case of patent foramen ovale plus ASD has been described (8). In only one of the above cases (14), was the atrial septal abnormality identified as an ostium primum defect.

The present report describes a case of congenital cardiac disease in a four week old male, Welsh pony foal in which postmortem evaluation led to a diagnosis of ostium primum defect, hypoplastic right ventricle and tricuspid dysplasia.

CASE HISTORY

The subject was a male, Welsh Mountain pony foal. The animal was well developed and the owner had noted no abnormality until the mare and foal were turned out to pasture. The foal exercised normally initially, but tired rapidly and showed respiratory distress.

The animal was first examined clinically at ten days of age at which time it was in good body condition and vigorously resisted restraint. A harsh, pansystolic, crescendo-decrescendo, grade IV/V murmur was auscultated on the left side of the thorax. The

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murmur radiated widely but was loudest anteriorly and dorsally at the third intercostal space at a level half way between the shoulder and the point of the elbow. There was a palpable thrill. On the right side a similar murmur could be heard anteriorly. At the time of examination the foal had a heart rate of 240 beats per minute and a respiratory rate of 60 breaths/minute. Respirations were labored and lung sounds were harsh and moist. Mucous membranes were cyanotic and the capillary refill time three seconds. Congenital cardiac disease with major right-to-left shunt was diagnosed. The foal was killed at four weeks of age and submitted for postmortem examination.

POSTMORTEM FINDINGS

Significant findings were restricted to the heart. On external inspection of the organ, the right ventricle appeared to be hypoplastic, the left ventricle dilated. The relationship of all major vessels appeared to be normal. A 1.5 cm diameter defect was noted in the cranial interatrial septum (Figs. 1 and 2). The foramen ovale was patent but was normal in structure and location. The septal cusp of the tricuspid valve was dysplastic (Fig. 1), being marked only by a narrow ridge of small, partially conjoined cystic structures approximately 3-5 mm in diameter and filled with a blood-stained fluid. The parietal cusp of the tricuspid valve was rudimentary and the tricuspid ostium was mildly hypoplastic (circumference 8 cm). The musculi papillaris parvi and musculus papillaris subarteriosus were absent. The right ventricle was hypoplastic. There was moderate muscular (infundibular) stenosis of the right heart outflow tract. The pulmonic valve was bicuspid and dysplastic, the valve cusps being thickened and irregular, and the pulmonary ostium was hypoplastic (circumference 3.75 cm) (Fig. 3). One centimetre above the pulmonic valve the pulmonary artery exhibited a discrete constriction beyond which the artery was dilated and thin-walled.

The left atrium and ventricle appeared moderately dilated, as did the left A-V and aortic valves (circumferences 10 and 7.5 cm respectively). There was no other evidence of left A-V insufficiency, but a deep cleft was present in the septal cusp of the mitral valve with prominent chordae tendineae adherent at their distal extremity to the base of the adjacent valve leaflet (Fig. 2). The cleft effectively divided the septal leaflet in two. Halfway between this cleft and the ostium primum was a small fossa in the otherwise smooth floor of the left atrium. The origin and distribution of the cranial and caudal vena cavae, the coronary arteries and the pulmonary veins were normal.

Postmortem findings were pulmonic valve dysplasia, infundibular, valvular and supravalvular pulmonic stenosis, hypoplastic right ventricle, tricuspid dysplasia and stenosis, persistent ostium primum, valvular-competent foramen ovale, dysplasia of the mitral valve and left ventricular dilatation and hypertrophy.

DISCUSSION

Patent foramen ovale must be differentiated from true atrial septal defect (ASD), in which septal tissue is deficient (9). Such defects may arise through the presence of an excessively large ostium secundum (fossa ovalis defect, ASD II), failure of the interatrial septum primum to form in the region of the sinus venosus (cranial caval defect), or caudal vena cava (caudal caval defect), or failure of the septum primum to completely partition the atria, leading to a persistent ostium primum (ASD I). The latter may represent a form of atrioventricular (A-V) canal defect, with incomplete fusion of the dorsal and ventral A-V endocardial cushions; ostium primum defects almost invariably occur in association with other A-V canal abnormalities. The interatrial septum may also fail to form at all (common atrium, cor triloculare biventriculare). The diagnosis of ostium primum defect made in this case was based upon the location of the defect and was supported by the presence of a normal, valvular-competent foramen ovale and a defect of the mitral valve.

The embryonic development of the interatrial septum in the horse has been described recently (16) and appears to follow the pattern found in man, though the various events occur up to five days earlier in the horse. By gestational age 22-25 days, the primitive common atrium is already partially divided by the septum primum (Fig. 4a,b) which originates in the roof of the common atrium and extends with its free margin towards the common A-V canal, joining the dorsal and ventral A-V endocardial cushions at its extremities. The free margin of the septum thus defines a communicating passage between the primitive left and right atria, the ostium primum.

While the ostium primum is still patent, and before the A-V endocardial cushions have fused, multiple fenestrations appear in the caudal por-

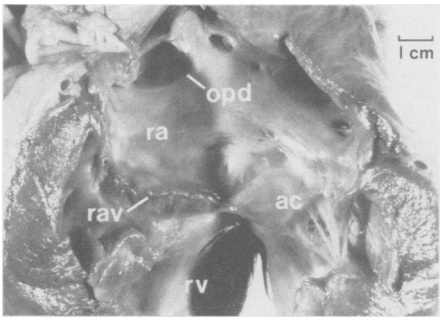

Fig. 1. Exposed view of the right atrium and ventricle. Ostium primum defect (opd). Right Atrium (ra). Dysplastic septal cusp of the right atrioventricular valve (rav). Anterior cusp of the right atrioventricular valve (ac). Hypoplastic right ventricle (rv).

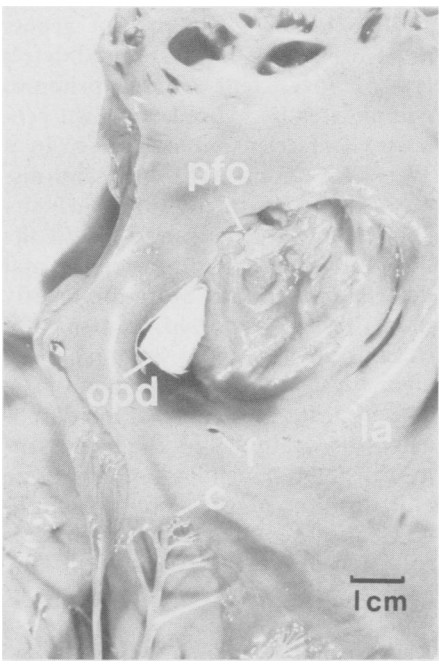

Fig. 2. Exposed view of the left atrium (la), and left atrioventricular valve. Patent foramen ovale (pfo). Ostium primum defect (opd). Fossa in the floor of the left atrium (f). Cleft in the septal cusp of the left atrioventricular valve (c).

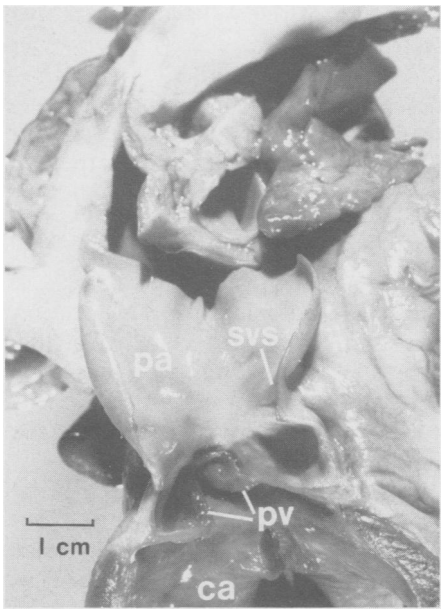

Fig. 3. Right heart outflow tract. Pulmonary artery (pa). Supravalvular stenosis (svs). Thickened, stenotic bicuspid pulmonic valve (pv). Conus arteriosus (ca).

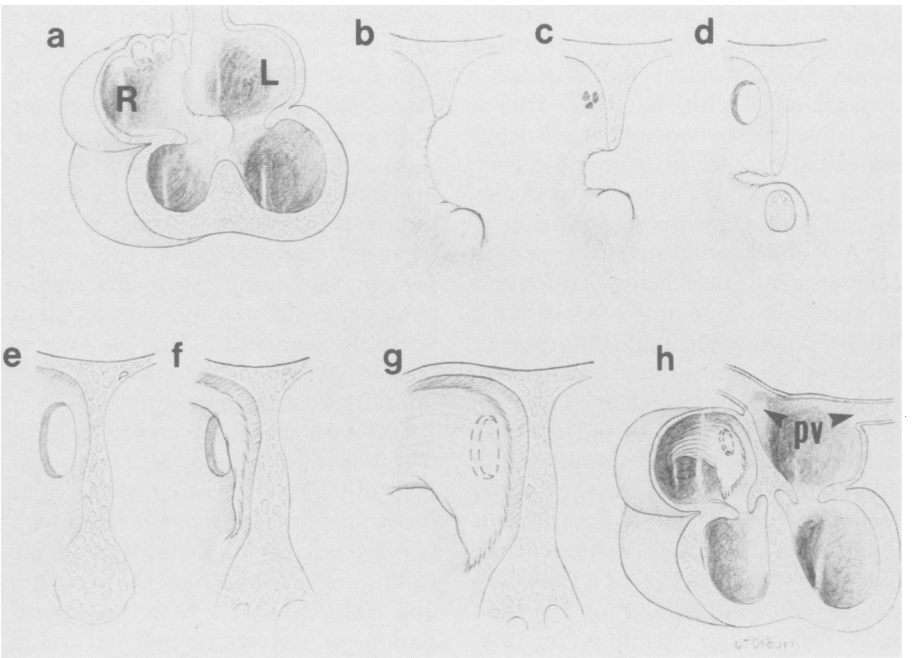

Fig. 4. Diagrammatic representation, embryogenesis of the interatrial septum. Left and right sides of the heart are marked. Pulmonary veins — pv. Refer to the text for explanation.

TABLE I. Recorded Cases of Congenital Cardiac Disease and Discontinuity of the Atrial Septum in the Horse[a]

		Reference									
Defect	Case	10	5	12	13	6	7	14	11	15	19	8
Patent foramen ovale			X			X	X					X
Common A-V canal								X				
Atrial septal defect	X	X		X	X				XX	XX	X	X
Atresia, right A-V valve		X	X			X			XX			
Dysplasia, right A-V valve	X											
Hypoplastic right ventricle	X		X			X	X		XX			
Hypoplastic left ventricle											X	
Pulmonary atresia				X	X		X					
Valvular pulmonic stenosis	X	X	X									
Supravalvular pulmonic stenosis	X		X									
Bicuspid pulmonic valve	X								X			
Dysplasia, left A-V valve	X								XX			
Ventricular septal defect		X	X	X	X	X			XX	XX		
Dextrarotation of the aorta		X			X							
Coronary artery anomaly			X									
Patent ductus arteriosus							X			XX	X	
Aortic arch anomaly										XX		
Truncus arteriosus communis				X								

[a]Atrial defects categorized according to present authors interpretation — see text

tion of the septum primum (Fig. 4c). By gestational age 26-28 days these have fused to form a large, distinct ostium secundum, while ostium primum is small but still visible (Fig. 4d). By days 30-32, the A-V endocardial cushions have fused and form the medial walls of the A-V ostia and ostium primum has closed (Fig. 4e). By days 33-35 the semilunar valve cusps are being formed by excavation and the primitive A-V valves are making their appearance. Septum secundum does not appear until days 45-49 at which point it is evident as primitive ridges located adjacent to septum primum in the right atrium (Fig. 4f). It develops from the craniodorsal side of the right atrium but never closes, instead forming a flap which covers ostium secundum, the concavity of its free margin facing the opening of the caudal vena cava (Fig. 4g, h). Septum secundum fuses partially with septum primum and creates a passageway or canal by which blood passes beneath its free edge and through ostium secundum into the left atrium. This passageway becomes the foramen ovale.

All of the structures which were found to be abnormal in the present case were formed before a gestational age of 40-45 days. The foramen ovale in this case was anatomically patent, which in a four week old foal may be regarded as normal (4) and was structurally normal. Septum secundum developed normally with the exception of its failure to form in the region of the persistent ostium primum. Atrial septal defect I need not be associated with abnormalities of the foramen ovale (9,17).

Persistence of ostium primum is rarely an isolated finding. The syndrome of persistent ostium primum together with cleft of the aortic (septal) cusp of the left A-V valve is well recog-

nized as one form of partial A-V canal abnormality in man (17). Though ostium primum defects may also occur in association with cleft of the tricuspid valve, other concurrent tricuspid abnormalities are also possible (17). These associations occur because closure of ostium primum and fusion of the A-V endocardial cushions are concurrent events, both being required for complete septation of the A-V canal; it has been suggested that abnormal or deficient growth of the endocardial cushions may be a primary factor in failure of ostium primum to close (18). In the present case the changes found in the left A-V valve and on the floor of the left atrium are consistent with this hypothesis. The malformation of the tricuspid valve was not of a type consistent with simple failure of the right A-V endocardial cushions to fuse, though such abnormalities of the valve have been seen in asociation with ASD I in man (17).

A search of the veterinary literature revealed 13 cases of discontinuity of the atrial septum in horses. These are summarized in Table I. All of the reported cases occurred in association with other cardiac defects. Five were associated with tricuspid atresia (5,6,10,13). In one (5), the atrial lesion is clearly described as a patent foramen ovale, while in two cases there were associated pulmonic valve abnormalities and a ventricular septal defect. Nine of the cases had ventricular septal defects and six had hypoplasia of the right ventricle. One case had an atrial defect in association with hypoplastic left ventricle and patent ductus arteriosus (19); the location described for the defect in this case would suggest that it was unique in being of the caudal caval type (9). In man, ostium primum defects are most frequently associated with ostium secundum defects and left superior vena cava (9), while pulmonic valve stenosis is a less frequent association. Nine of the equine cases reviewed had some degree of obstruction to the flow of blood through the right heart.

With the exception of one reference to a bicuspid right A-V valve in a donkey (14) and reference to three unspecified cases of A-V defect (2), dysplasia of the tricuspid valve in the horse is not reported in the literature and the frequency of this abnormality cannot be assessed. However, in man dysplasia of the tricuspid valve is uncommon, though when found it is frequently associated with pulmonary valvular obstruction and enlargement of the right atrium and ventricle (20). In the present case, in addition to the tricuspid dysplasia, mild hypoplasia of the tricuspid and pulmonic ostia were present. This combination of defects is consistent with the hypoplastic right ventricle also found, and in man is almost always associated with an interatrial communication (9,21).

Our case shows the closest parallel with a form of pulmonary stenosis in man in which narrowing of the right heart outflow tract is associated with varying degrees of hypoplasia of the right ventricle and right A-V ostium and with ASD (16). With this combination of defects, blood returning from the systemic circulation bypasses the right heart by flowing through the ASD directly into the left atrium. This direction of flow is encouraged by elevated right atrial pressures secondary to increased resistance to right heart outflow and the low capacity of the right ventricle. The resultant right-to-left shunt causes systemic hypoxia and cyanosis. Flow of blood through the lungs for oxygenation is achieved via 1) very limited flow via the right ventricle and pulmonary artery and 2) increased flow through the bronchial arteries, which have been shown to be capable of supplying blood directly to the alveolar capillary network in the horse (22). Patency of the ductus arteriosus would be of some value in increasing systemic oxygenation since the shunt would be left to right. The ductus arteriosus was not patent in the present case.

A review of the literature revealed only one previously identified case of persistent ostimum primum in the horse (14). However, examination of four of the reported cases of ASD suggest that these were probably ostium primum defects. In the first report (8), two defects were described; "... a large opening was found in the fenestrae of the fossa ovalis and an opening cranial to the limbus fossa ovalis (the concave lateral margin)." Though the illustration accompanying the article is not clearly marked, the location of the defect indicates that this was probably a persistent ostium primum. The condition of the A-V valves was not stated.

In the second report (11), ASD is described in two Thoroughbreds, both three months of age, in association with tricuspid atresia, right ventricular hypoplasia, left ventricular hypertrophy and dilatation and large subaortic ventricular septal defect. In addition one of the animals had a bicuspid pulmonic valve. The location of the atrial septal defect in the illustration is most consistent with persistent ostium primum, rather than patent foramen ovale as suggested by the authors.

In the third report (6), atresia of the tricuspid valve together with hypoplastic right ventricle and ventricular septal defect is again described as accompanying a discontinuity of the atrial septum. The defect (designated as a patent foramen ovale by the author), is clearly shown to be located in the cranial portion of the atrial septum, a location far more consistent with an ostium primum defect. The associated presence of tricuspid atresia and the anomalous attachment of an A-V valve cusp to the margins of the interatrial defect in this case also suggest an A-V endocardial cushion defect. Ostium primum defects may occur more commonly in the spectrum of congenital cardiac disease in the horse than a cursory review of the literature indicates.

The clinical significance of uncomplicated atrial septal defects is limited. The pressure gradient between left and right atria results in a left-to-right shunt. Over a number of years the resultant volume overload on the right heart and increased pulmonary circulation will lead to pulmonary hypertension and right-sided hypertrophy. In the case of a large defect, right-sided failure and elevation of central venous pressure causing reversal of the shunt and cyanosis is possible. The likelihood of this occurring will depend not only upon the size of the defect but also upon other modifying factors such as respiratory disease. In complicated cases the consequences will depend upon the nature of the additional defects. Since, with the exception of abnormalities of the mitral valve in cases of ASD involving the A-V canal, atrial septal defects in the horse usually appear to be associated

with various degrees of obstruction to the flow of blood through the right heart and its outflow tract, there is likely to be sufficient elevation of pressure in the right atrium to cause a right-to-left shunt at the level of the atria. The attendant problems of systemic hypoxia and underperfusion of the lungs result in clinical cyanosis, especially at exercise, and limited exercise tolerance. Even in the absence of further complications such as bacterial endocarditis, a limited lifespan may be anticipated.

Since the completion of this paper a second foal by the same sire has been delivered. This foal shows very similar clinical signs to the animal described herein, but was unavailable for postmortem evaluation.

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